# PERICARDIAL DISEASE: WHAT THE GENERAL CARDIOLOGIST NEEDS TO KNOW



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*Heart* 2007;**93**:993–1000. doi: 10.1136/hrt.2005.086587

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Pericardial disease remains an important cause of morbidity and mortality, spanning a complex spectrum from asymptomatic and transient to severely symptomatic and life threatening. Knowledge about the presenting symptoms, clinical findings, diagnosis and management is essential for effective clinical management.

This article focuses on the pertinent features of the major pericardial diseases seen in clinical practice and the recommended diagnostic and treatment strategies. In many cases the management of pericardial disease is based on best clinical practice; however, evidence based recommendations are emerging. Published data, gleaned from observing numbers of patients, must be crystallised into an individualised management plan.

## THE NORMAL PERICARDIUM

The pericardium consists of two layers that surround the heart and the proximal aorta, pulmonary artery, vena cava and pulmonary veins. The thick fibrous outer layer, which attaches to the adventia of the major vessels, diaphragm, sternum and vertebrae, provides strong structural support for the heart.[1] The inner layer is a thin serous membrane composed of only a single row of mesothelial cells which lies both on the surface of the heart, where it is called the visceral pericardium, and by folding back on itself and underlying the fibrous layer, forms the lining of the parietal pericardium. Normally the pericardium is <2 mm thick. The pericardial space is a blind sac contained within the visceral and parietal pericardium and usually contains only a small amount of pericardial fluid. The transverse sinus is the part of the pericardial sac which lies between the great vessels and the oblique sinus lies posteriorly between the pulmonary veins.

Pericardial fluid is produced by the visceral pericardial cells and resembles an ultrafiltrate of plasma that serves to lubricate and reduce friction between the visceral and parietal pericardial surfaces. The pericardial fluid is drained by the thoracic and right lymphatic ducts. Normally there is between 10–50 ml of pericardial fluid.

The pericardium, although not critical for survival, has several important functions including: limiting acute dilatation of the heart and excessive cardiac motion within the chest; optimising cardiac shape; balancing right and left sided ventricular outputs via diastolic and systolic interactions (ventricular coupling); and preventing spread of adjacent infection or neoplasia to the heart.[1]

Being a relatively inelastic and non-compliant structure, the pericardium limits the total volume of the contained cardiac chambers and pericardial fluid, which is referred to as pericardial constraint. It is the parietal pericardium, more than the visceral pericardium, that mediates most of this effect. Under normal conditions, this constraint is not particularly pronounced.

Pericardial pressure is normally low and follows intrathoracic (or intrapleural) pressure and right atrial pressure. It is influenced by respiration.

## ABNORMAL PERICARDIAL PHYSIOLOGY

Raised intrapericardial pressure can occur by three main mechanisms: (1) increased fluid within the intrapericardial space; (2) increased volume of the cardiac chambers; or (3) increased stiffness of the pericardium. Raised intrapericardial pressure has three potential adverse effects on the heart: (1) a compressive effect which limits diastolic filling of the heart; (2) increased diastolic filling pressures; and (3) reduced stroke volume and cardiac output. Compensatory mechanisms are activated, but severe elevation of pericardial pressure can rapidly lead to death if not treated.

The concept of pericardial compliance is integral to understanding the clinical effects of an accumulating pericardial effusion. In the normal situation, the pericardium has a small capacitance reserve that will accommodate only small increases in cardiac chamber size and/or pericardial fluid volume of about 150–250 ml before significant increases in pericardial pressure occur.[1] Once the

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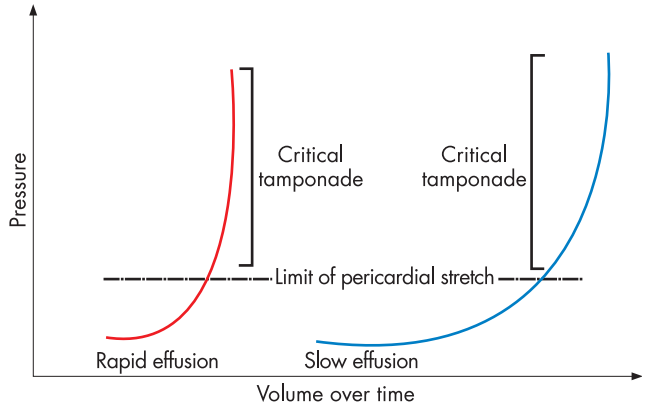


**Figure 1** Cardiac tamponade. Pericardial pressure–volume (or strain–stress) curves are shown in which the volume increases slowly or rapidly over time. In the left panel, rapidly increasing pericardial fluid first reaches the limit of the pericardial reserve volume (the initial flat segment) and then quickly exceeds the limit of parietal pericardial stretch, causing a steep rise in pressure, which becomes even steeper as smaller increments in fluid cause a disproportionate increase in the pericardial pressure. In the right panel, a slower rate of pericardial filling takes longer to exceed the limit of pericardial stretch, because there is more time for the pericardium to stretch and for compensatory mechanisms to become activated. Reproduced with permission from Spodick D. Acute cardiac tamponade. *New Engl J Med* 2003;**349**:684–90. Copyright 2003 Massachusetts Medical Society. All rights reserved.

pericardial capacitance reserve is reached, any further increase in contained volume dramatically increases pericardial pressure as this occurs on the steep incline part of the pressure–volume curve (fig 1).

## ACUTE PERICARDITIS

Acute pericarditis is a disorder characterised by acute inflammation of the pericardium. There are many different causes (webtable 1[2–10]) (to view webtable 1 visit the *Heart* website—http://heart.bmj.com/supplemental). Primary acute pericarditis, without an obvious underlying cause, is presumably of viral aetiology. Most secondary causes of acute pericarditis are evident before pericardial involvement occurs.

Presentation is usually with pleuritic chest pain in a haemodynamically stable patient. There may be a history of a recent viral illness or another predisposing condition. The duration of symptoms is usually short, and the pain is typically sharp, worse on deep inspiration, coughing and on lying flat. The pain can be intermittent or constant. It can be severe enough for the patient to seek urgent medical assessment. Other symptoms that may be present include a dry cough, dyspnoea, malaise, myalgia and fever.

Examination findings are listed in webtable 2[2 4 8 11–23] and usually reveal a stable patient who is normotensive (to view webtable 2 visit the *Heart* website—http://heart.bmj.com/supplemental). A pericardial friction rub may be audible on auscultation in approximately 35% of patients, although it is often transient.[13 14] Some believe it is more likely in those without an effusion, although this has been debated. It is felt to be caused by the two inflamed layers of pericardium moving against each other in atrial systole, ventricular systole and/or rapid diastolic filling. Sinus tachycardia and fever may be present.

Investigations that should be performed and the typical ECG findings are outlined in webtable 2. Elevated cardiac enzymes are not uncommon in acute pericarditis and usually do not imply ischaemic heart disease. In one study of 69 patients with acute pericarditis, cardiac troponin I was detectable in 49% and above 1.5 ng/ml$^{-1}$ in 22%.[24] Approximately half the patients with elevated troponin I levels had coronary angiography performed and no coronary artery disease was detected.[24] Elevated troponin values were felt to reflect superficial myocardial inflammation and were not an adverse prognostic marker after a mean follow-up of 24 months.[25]

Echocardiography should be performed in all patients with pericarditis, for a number of reasons, and has been given a class 1 indication according to the 2003 taskforce of the American College of Cardiology, the American Heart Association and the American Society of Echocardiography. It is helpful in detecting pericardial effusions that may be present in up to 66% of patients and this finding is, in general, supportive of the diagnosis.[14] Echocardiography is also helpful in looking for other complications and for evaluating other differential diagnoses. Regional wall motion abnormalities or global dysfunction may be present and may reflect either myocarditis, or ischaemia/infarction secondary to underlying coronary disease. Echocardiography may also detect pericardial thickening or suggest an alternative cardiac diagnosis.

The diagnosis of acute pericarditis is made by clinical assessment in combination with supportive examination and ECG findings. Many authors recommend two out of the three following criteria for a firm diagnosis: typical pleuritic chest pain; pericardial friction rub; and widespread ST segment elevation on the ECG.[2 14]

Treatment is symptomatic and consists of anti-inflammatory medication, predominantly non-steroidal anti-inflammatory drugs (NSAIDs) with or without colchicine. Aspirin is the preferred choice at a dose range of 325–650 mg orally four times daily.[14] Ibuprofen is the next recommended NSAID due to its low side effects, favourable impact on coronary blood flow, and large dose range of 300–800 mg, 6–8 hourly.[15] NSAIDs can be continued until any effusion and/or symptoms have resolved (generally for days to weeks), with the dose being reduced as clinically indicated.[15] Many uncomplicated episodes of acute idiopathic pericarditis only require 1–4 days of treatment, although the optimal duration of treatment recommended is unknown. Gastrointestinal protection is recommended for all patients.[13–15] Colchicine, 500 μg daily, can be given as monotherapy or in combination with an NSAID for symptomatic relief and for the prevention of further episodes.[14] Oral steroids are usually reserved for patients with underlying connective tissue disease, uraemia or recurrent disease according to the European Society of Cardiology.[15] As these are weaned, colchicine or NSAID treatment is usually introduced. Steroid use should be restricted as it is an independent risk factor for recurrence.[14]

The COPE trial provides randomised data that further supports and expands the role of colchicine in the primary treatment of acute pericarditis.[14] Of 120 patients recruited, 60 received aspirin for 1 month and 60 received combination treatment with aspirin for 1 month and colchicine for 3 months. Results showed a statistically significant reduction in the pericarditis recurrence rate of 32.3% vs 10.7% with

combination treatment ($p = 0.004$, number needed to treat = 5). Colchicine also significantly reduced symptom persistence at 72 h (11.7% vs 36.7%, $p = 0.003$). Colchicine should therefore be considered in the primary treatment of acute pericarditis.

Diagnostic pericardiocentesis has a low yield in the setting of acute pericarditis and is not routinely recommended unless there is a high clinical suspicion of purulent pericarditis.[2–4] Therapeutic pericardiocentesis is reserved for those with pericardial effusion and tamponade, or a significant effusion with a poor clinical course after 7–10 days of NSAID treatment, although the latter is controversial.[2]

Many patients with acute pericarditis do not require hospitalisation and can be followed up after appropriate evaluation as an outpatient. In a recent review of 300 patients, 254 (85%) were managed as outpatients with NSAIDs, if they did not have high risk features on initial clinical and echocardiographic assessment.[13] These high risk features were fever >38℃, subacute onset, immunosuppression, trauma, oral anticoagulant treatment, myopericarditis, large pericardial effusion (>2 cm) or tamponade. Outpatient treatment was efficacious in 87% of cases. During the study, 13% of patients were admitted because of failure of NSAIDs to control symptoms. Higher rates of recurrent disease and constriction were seen in NSAID resistant patients. In those treated as an outpatient, there was no incidence of serious complications after a mean follow-up of 38 months.

Specific types of acute pericarditis may also require additional treatment, such as antimicrobial therapy and surgical drainage for bacterial pericarditis, antituberculous treatment for tuberculosis infection, and haemodialysis for uraemic pericarditis. These are discussed in further details in the reference texts.[1 15]

After an episode of acute pericarditis, all patients require follow up assessment for complications such as recurrent disease and constriction.

## PERICARDIAL EFFUSION

A pericardial effusion is present when there is increased fluid within the pericardial space. This fluid can be serous, serosanguinous, pus, lymph or blood. There are many different possible causes (webtable 1). The reported aetiology varies with the study population and the diagnostic criteria and exclusions used. Routine diagnostic pericardiocentesis has a low yield, particularly in smaller effusions. In large effusions, the diagnostic yield of pericardiocentesis has been reported as between 7–26% and is higher in cardiac tamponade.[2–6 26] This supports the notion that routine diagnostic pericardiocentesis in the absence of cardiac tamponade is not indicated. Therapeutic pericardiocentesis may, however, be required for symptoms and/or cardiac tamponade.

The relationship between effusion size and adverse effects is influenced by the time course of accumulation. Many effusions are small, only discovered incidentally and are asymptomatic. Larger effusions that collect slowly may initially be asymptomatic, but cause dyspnoea or tamponade at a late stage. Rapidly accumulating effusions can result in cardiac tamponade and death, even when relatively modest in size. In this instance the pericardial pressure can rise rapidly because there is insufficient time for the non-compliant pericardium to stretch (fig 1).

Diagnosis of a pericardial effusion is usually achieved by echocardiography and can be circumferential or localised (loculated). Pericardial effusions are usually classified as small (<1 cm), moderate (1–2 cm) or large (>2 cm). The assessment of haemodynamic effect will be discussed in the next section.

The management of small or moderate pericardial effusions, without tamponade, is usually conservative, with clinical and echocardiographic surveillance. If the pericardial effusion is likely to be purulent then it should be drained. If the effusion is felt to be malignant, pericardiocentesis is recommended if confirmation would change management and can be performed safely.

The management of large, idiopathic, chronic pericardial effusions (asymptomatic and present for more than 3 months), without cardiac tamponade, is controversial. Many believe they should be monitored and managed conservatively, unless cardiac tamponade develops, and that they overall have a good prognosis. Merce *et al* reviewed 71 patients with large pericardial effusions in the absence of tamponade or a suspicion of purulent disease and found a relatively benign course.[26] Another study by Sagrista-Sauleda, however, showed a reasonably high risk of the development of overt tamponade and recommended a more invasive approach.[27] Pericardiocentesis will resolve the effusion in a proportion of cases. A risk–benefit assessment needs to be made for each patient, weighing the risk of pericardiocentesis against potential diagnostic and therapeutic benefit.

Pericardial effusions in patients with known malignancy can be neoplastic, idiopathic, or due to radiation, drugs or other conditions. Pericardiocentesis with analysis of cytological fluid is usually positive in 65–85% of patients with malignant effusions.[28] Open subxiphoid pericardial biopsy may diagnose many of the remaining patients. The management of large symptomatic malignant effusions, with or without tamponade, is usually by pericardiocentesis which had a success rate of 97% and a complication rate of less than 3%.[28] The risk of recurrence varies and is reduced by prolonged initial catheter drainage, and by the addition of systemic chemotherapy and/or radiotherapy in many cases.[28] The use of intrapericardial sclerosing agents and intrapericardial chemotherapy has been limited by the side effects of chest pain and atrial arrhythmias. Options for recurrent malignant effusions include repeat pericardiocentesis, subxiphoid window, pleuropericardial window and thoracotomy with pericardectomy.[28] A percutaneous technique has also been used to create a pericardial window.[28] The overall survival in patients with malignant pericardial effusions is poor and generally in the order of 10–13 months with breast cancer and less than 6 months in patients with other cancers. Survival is worse in patients with positive cytology in the fluid.

The risk of significant pericardial effusion/haematoma requiring treatment, after cardiac surgery, is approximately 1%. The effusion is often localised and the typical echocardiographic findings of tamponade may not be present. Transoesophageal imaging may be required for diagnosis if transthoracic images are suboptimal. If unexplained clinical deterioration occurs in the presence of a significant pericardial effusion/haematoma postoperatively, surgical exploration should be strongly considered even in the absence of echocardiographic markers of tamponade. Postoperative effusion/haematoma with tamponade can be treated with

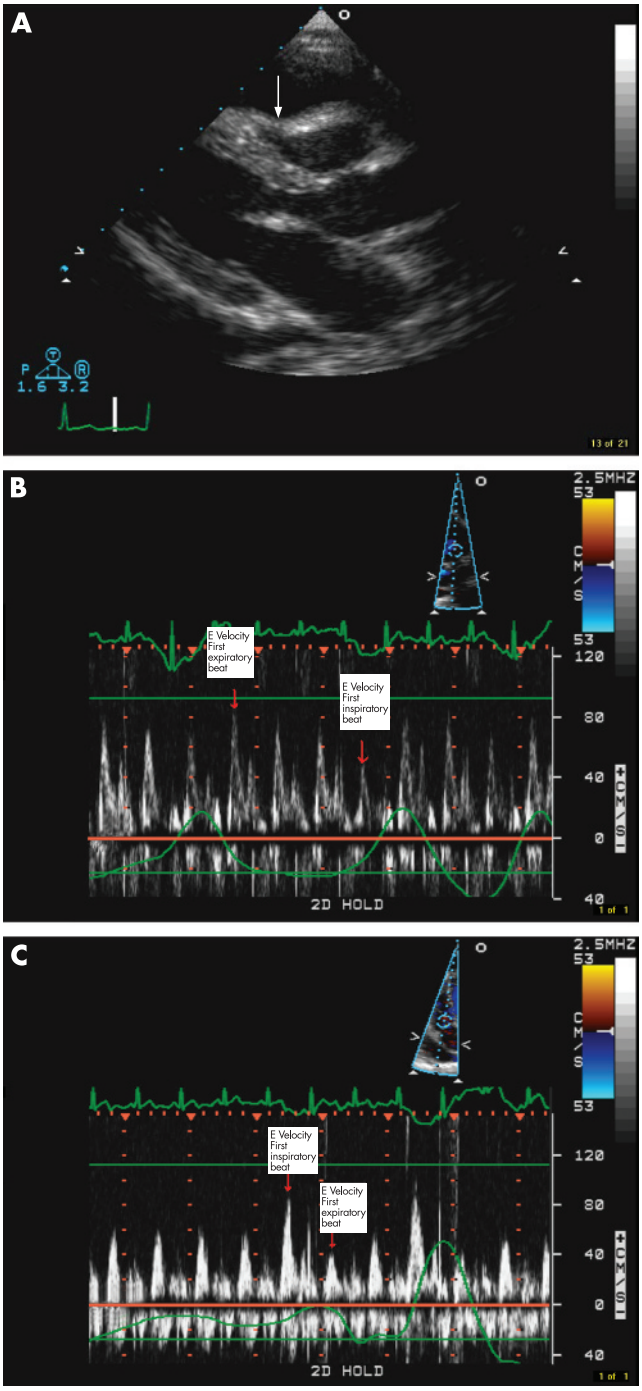


**Figure 2** (A) Transthoracic parasternal long axis echocardiographic image showing a large pericardial effusion. Arrow points to diastolic right ventricular compression. (B and C) Pulsed wave Doppler of the mitral and tricuspid valves showing increased respiratory variation in the same patient in panel A, who clinically had signs of cardiac tamponade.

percutaneous or surgical drainage, depending on local experience. Surgical drainage should be particularly considered if there is active bleeding or localised posterior pockets.

## CARDIAC TAMPONADE

Cardiac tamponade is a life threatening condition that is diagnosed clinically by elevated jugular venous pressure, hypotension and pulsus paradoxus in the setting of a pericardial effusion. It occurs when an accumulation of fluid within the pericardial space causes a significant rise in intrapericardial pressure. When intrapericardial pressure becomes higher than intracavitary pressure, compression of the heart occurs with reduced diastolic filling. This is first seen on the right side, rather than the left, due to the thinner walls and reduced chamber pressure. For similar reasons, the right atrium usually is compressed before the right ventricle. If the effusion is localised, compression of a single chamber can occur. If diastolic compression becomes too significant to be overcome by elevated filling pressures and other compensatory mechanisms, cardiac output and blood pressure will fall.

Many of the clinical and echocardiographic features are explained by the above findings and the effect of respiration on intracardiac flows and pressures.

The typical causes, and clinical and echocardiographic findings, are shown in webtables 1 and 2 and fig 2. Treatment is with prompt drainage of pericardial fluid (see Pericardiocentesis key points).

## CONSTRICTIVE PERICARDITIS

Constrictive pericarditis is usually characterised by a thickened, adherent pericardium that restricts ventricular filling and limits chamber expansion and maximal diastolic volumes. Elevated filling pressures are required to maintain adequate cardiac output. End-diastolic pressures in all heart chambers are usually elevated and equalised. Compensatory mechanisms are activated but may ultimately fail, leading to elevated venous pressure, oedema and diminished cardiac output.

The aetiology of pericardial constriction is changing (webtable 1).

Clinical findings are listed in webtable 2 and the presentation is usually with features mimicking right-sided heart failure with low cardiac output. Typical symptoms are exertional dyspnoea, peripheral oedema, abdominal distension and fatigue. Pulmonary oedema is not normally a feature.

Diagnosis is usually made by a combination of clinical findings, chest *x* ray, echocardiography, CT or MRI imaging and in many cases invasive haemodynamic studies (webtable 2, fig 3).

Echocardiography is an important tool for suggesting or supporting the diagnosis. The typical findings are listed in webtable 2.

In patients with notably elevated left atrial pressure, respiratory variation of the Doppler inflows may not be present unless preload is reduced by head-up tilt or diuretics.[29]

Not all patients with proven constriction at surgery actually have increased respiratory variation of Doppler inflows. Of 19 patients with surgically confirmed constrictive pericarditis, for example, nine (47%) did not have more than 25% respiratory variation of mitral E wave velocity.[30]

In patients with atrial fibrillation, changes in mitral E velocity may reflect variations in ventricular filling rather than constriction.

Patients with chronic obstructive pulmonary disease (COPD) without constriction may have increased respiratory variation in mitral and tricuspid inflow velocities and this may occasionally cause diagnostic difficulty. In contrast to constriction, variation is not typically maximal on the first inspiratory beat. In addition, mitral inflow is less restrictive with lower E/A

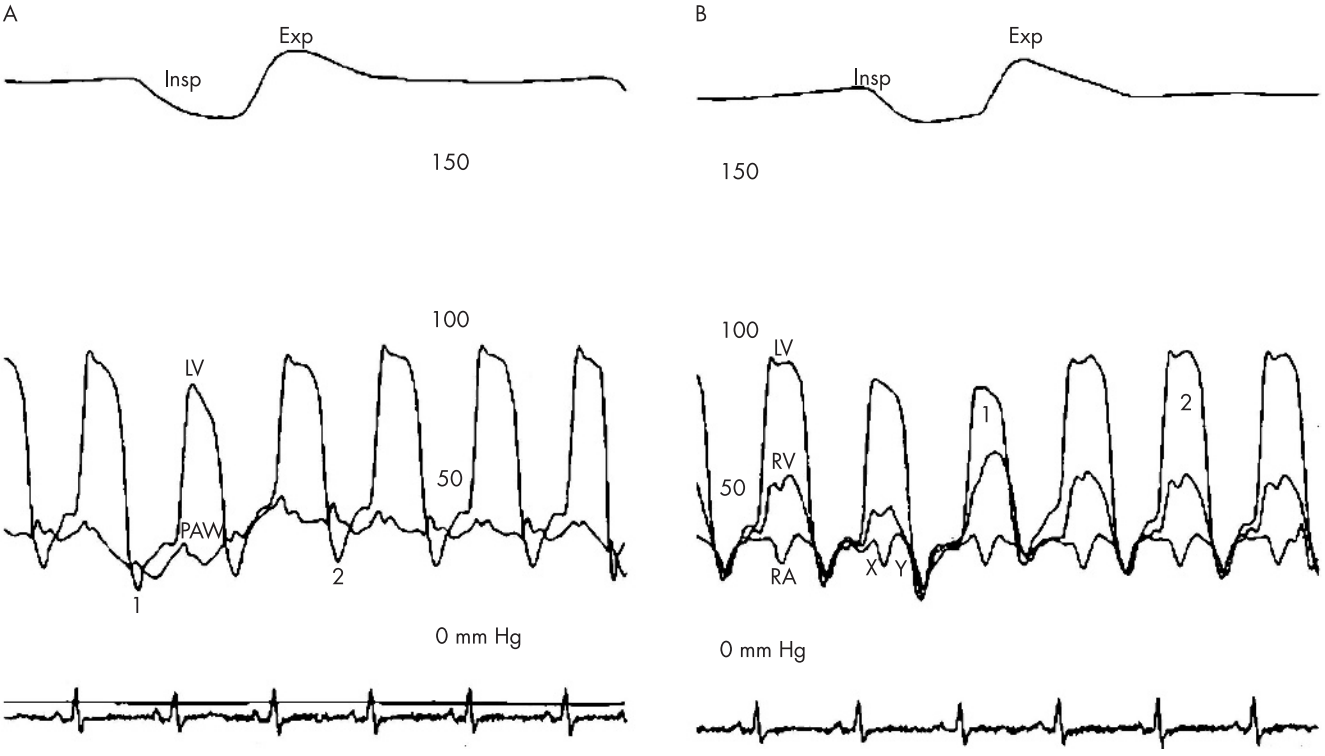


**Figure 3** (A) Simultaneous recordings of left ventricular (LV) and pulmonary capillary wedge (PAW) pressures demonstrating dissociation of intrathoracic and intracardiac pressures. Note the fall in early diastolic gradient with inspiration (beat 1) and the rise with expiration (beat 2). The nasal respirometer tracing is also shown. (B) Simultaneous recordings of left ventricular (LV), right ventricular (RV) and right atrial (RA) pressures in same patient, again demonstrating ventricular interdependence. Note the discordance in the left and right ventricular systolic pressures with respiration. There is a pronounced rise in right ventricular pressure during inspiration (beat 1), during which time the left ventricular pressure is falling. Note the rapid X and Y descents in the right atrial tracing. The nasal respirometer tracing is also shown. Exp, expiration; Insp, inspiration. Reproduced with permission from Higano ST, Azrak E, Tahirkheli NK, *et al*. Hemodynamic rounds series 2: hemodynamics of constrictive physiology: influence of respiratory dynamics on ventricular pressures. *Catheter Cardiovasc Interv* 1999;**46**:473–86. Global Rights Dept, John Wiley and Sons, Inc.

ratios than in patients with constriction and longer deceleration times.[31] Analysis of the superior vena cava (SVC) Doppler signal may be useful in this setting. SVC Doppler shows pronounced increases in forward flow with inspiration in patients with COPD which is not usually seen in constriction (mean (SD) respiration variation in COPD of SVC systolic flow velocity 39.5 (18.8) cm/s vs 4.2 (3.4) cm/s in constriction).[31] In one study, 95% of patients with COPD had respiratory variation of SVC systolic forward velocity >20 cm/s or a >35% increase with inspiration.[31] In addition, COPD patients usually have systolic dominant superior vena caval Doppler flow rather than diastolic dominant in constriction.[31]

Evaluation of the pericardium by chest radiography and pericardial thickness by CT or MRI provides important supplementary information that often helps in making this diagnosis in the majority of affected patients. Invasive haemodynamic evaluation is particularly important in patients with suggestive clinical features and equivocal non-invasive tests. Typical catheterisation findings are described in webtable 2 and table 1, and in fig 3.

B-type natriuretic peptide (BNP) is usually normal or only mildly raised in constrictive pericarditis (128 (52.7) pg/ml) but significantly higher in patients with restrictive cardiomyopathy (825.8 (172.2) pg/ml) ($p<0.001$).[23] A more recent study, however, revealed that a significant difference in BNP values existed only between those patients with idiopathic constrictive pericarditis and patients with restrictive cardiomyopathy. No statistically significant difference was found between patients with secondary constrictive pericarditis (for example, secondary to radiation or post-cardiac surgery) and restrictive cardiomyopathy.[32] This may reflect co-existent cardiac muscle disease in patients with secondary constrictive pericarditis where certain disease processes may affect both pericardium and cardiac muscle.

Features that would support a diagnosis of restrictive cardiomyopathy rather than constrictive physiology are listed in table 1.

Treatment of established chronic constrictive pericarditis is by radical pericardectomy.

## TRANSIENT CONSTRICTION

Transient pericardial constriction has been reported in the following settings: acute pericarditis, cardiac surgery, collagen vascular disease, malignancy, tuberculosis, trauma and idiopathic. Typical patient characteristics and treatment modalities are discussed in detail elsewhere. The mechanism of transient constriction is felt to be secondary to an inflammatory mediated reduction in pericardial compliance; a trial of medical treatment may be appropriate in haemodynamically stable patients with recovery expected at an average of 3 months. It is not clear from the available evidence if medical treatment affects the natural history of this disorder. Treatment should be

**Table 1** Comparison of constrictive pericarditis and restrictive cardiomyopathy

| | Constrictive pericarditis | Restrictive cardiomyopathy |
|---|---|---|
| Most common aetiology | Idiopathic<br>Post-cardiac surgery<br>Radiotherapy<br>Connective tissue disease<br>Infection/tuberculosis<br>Malignancy | Idiopathic<br>Infiltrative; amyloidosis, eosinophilic diseases<br>Radiotherapy |
| Clinical symptoms | Dyspnoea, fatigue, peripheral oedema, ascites | Dyspnoea, fatigue<br>Peripheral oedema |
| Pulsus paradoxus | Uncommon | Absent |
| JVP height | Elevated | Elevated |
| JVP waveform | Prominent X and Y descents | Prominent X descent |
| Kussmaul's sign | Often present | Absent |
| Pericardial knock | Often present | Absent |
| ECG | | Low voltage QRS complexes in amyloidosis |
| Atrial size | Usually normal | Dilated |
| LV and RV size/systolic function/2D appearance | Usually normal | Normal or mildly reduced LV systolic function |
| Increased LV wall thickness | Absent | May be present in infiltrative causes |
| Septal motion | Abnormal | Normal |
| Septal position | Respiratory variation | Normal |
| Mitral/tricuspid regurgitation | Infrequent | Frequent (TR>MR) |
| Mitral inflow pattern PW Doppler | Increased E velocity<br>Shortened deceleration time<br>Mitral E/A ratio often >2.0 | Increased E velocity<br>Restrictive filling pattern<br>Mitral E/A ratio >2.0, shortened DT (<160 ms) |
| Respiratory variation | Exaggerated and reciprocal respiratory changes in Doppler inflows; increased right sided Doppler velocities (tricuspid, pulmonary valve) and reduced left sided Doppler velocities (mitral, aortic) with inspiration. Opposite changes occur with expiration.<br>Percentage change from expiration to inspiration: †mitral E velocity >−25% (−33 (9)%), aortic velocity −14 (5)%, tricuspid E velocity 44 (22)%, pulmonary artery velocity 16 (4%)[19] | Normal with minimal respiratory variation of Doppler inflows<br>Percentage change from first beat of expiration to first beat of inspiration: †mitral E velocity −3% (4)%, tricuspid E velocity 17 (16)%, aortic velocity −4 (5)%, pulmonary artery velocity 5% (7)%[19] |
| IVRT | Duration increases from expiration to inspiration, increased respiratory variation IVRT >25% (50 (14)%)†[19] | Constant throughout respiration, minimal respiratory variation of IVRT duration 4 (7)%†[19] |
| Pulmonary vein PW Doppler | Increased respiratory variation of diastolic pulmonary vein Doppler flow >18%[20] | No significant respiratory variation |
| IVC | Dilated IVC<br><50% reduction in IVC width with inspiration | Dilated IVC<br><50% reduction in IVC width with inspiration |
| Hepatic vein | Prominent diastolic flow reversals in expiration | Prominent diastolic atrial flow reversals in inspiration |
| SVC Doppler | Diastolic prominence of forward flow (reduced systolic forward flow) | Diastolic prominence of forward flow (reduced systolic forward flow) |
| Pulmonary hypertension | Rare and mild | Frequent and moderate-severe elevation |
| Colour M mode propagation velocity (first aliasing contour) | Normal to increased >100 cm/s[20] | Reduced (<100 cm/s)[20] |
| Tissue Doppler; mitral annulus | Normal to increased E' velocity >8 cm/s[20] | Reduced E' velocity <8 cm/s[20] |
| CXR | Pericardial calcification | No pericardial calcification |
| Pericardium thickness on CT/MRI imaging | Increased | Normal |
| BNP | Normal to minimally elevated 128 (53) pg/ml[23] | Notably elevated 826 (172) pg/ml[23] |
| Catheterisation haemodynamics | Elevation and near equalisation of all diastolic pressures*<br>LVEDP-RVEDP ⩽5 mm Hg*<br>PASP ⩽55 mm Hg*<br>RVEDP/RVSP >1/3*<br>Dissociation of intracardiac and intrathoracic pressures with increased respiratory variation between the PCWP and early diastolic LV pressure gradient ⩾5 mm Hg[22] (fig 3A)<br>Increased ventricular interdependence; discordant changes of LV systolic pressure and RV systolic pressure with respiration[22] (fig 3B) | Elevation and near equalisation of all diastolic pressures*<br>LVEDP-RVEDP >5 mmHg*<br>PASP >40 mm Hg*<br>RVEDP/RVSP <1/3* |
| Endomyocardial biopsy | Usually normal myocardium | Abnormal or non-specific |
| Treatment | Pericardectomy | Medical treatment<br>± treatment of underlying disorder<br>± cardiac transplantation |

BNP, B-type natriuretic peptide; CT, computed tomography; CXR, chest *x* ray; ECG, electrocardiogram; IVC, inferior vena cava; IVRT, left ventricular isovolumic relaxation time; JVP, jugular venous pressure; LV, left ventricle; LVEDP, left ventricular end-diastolic pressure; MR, mitral regurgitation; MRI, magnetic resonance imaging; PASP, pulmonary artery systolic pressure; PCWP, pulmonary capillary wedge pressure; PW, pulsed wave; RV, right ventricle; RVEDP, right ventricular end-diastolic pressure; RVSP, right ventricular systolic pressure; SVC, superior vena cava; TR, tricuspid regurgitation.
This table presents an outline of parameters which can be useful in differentiating the above two conditions, but in the individual patient discordant data can occur, and the overall distinction should be based on an overall assessment.
*Suboptimal sensitivity and specificity of these criteria may limit the clinical usefulness in individual patients.
†Percentage change from the first beat of expiration to the first beat of inspiration. Values are presented as mean (SD). All p values are <0.05 comparing percentage respiratory change in constrictive pericarditis versus restrictive cardiomyopathy and constrictive pericarditis versus normal.[19] The respiratory variation (%) represents a continuum and cut-offs are not absolute; −% indicates a negative direction of change.

directed at symptoms while monitoring for clinical and echocardiographic improvement. If deterioration occurs, early surgery should be considered.

## EFFUSIVE CONSTRICTIVE PERICARDITIS

Effusive constrictive pericarditis is a condition where a pericardial effusion coexists with constriction of the heart by the visceral layer of the pericardium. The pericardial effusion is usually tense and causes pericardial tamponade. The clinical diagnosis rests on the demonstration that a clinical and haemodynamic picture of constriction persists after drainage. Simultaneous pericardiocentesis and right heart catheterisation will show a persistent elevation of right atrial pressure despite the normalisation of intrapericardial pressure following drainage. A study by Sagrista-Sauleda *et al* identified 15 patients who met the criteria for effusive constrictive pericarditis who were followed for a median period of 7 years.[9] All 15 patients had clinical manifestations of right heart failure. The underlying aetiology is shown in webtable 1. Pericardectomy was required by seven patients who had persistent constriction and clinical features of severe and persistent heart failure who were operative candidates. Significant thickening and adhesions were found of both the visceral and parietal pericardial layers in all these patients at surgery. Spontaneous resolution occurred in three patients.

## MIXED CONSTRICTION AND RESTRICTION

Mixed constriction and restriction is a disorder characterised by difficulty in making and confirming the diagnosis, less clearly defined treatment options and in general, a poorer outcome. This diagnosis can be suspected on cardiac imaging using three criteria: (1) localised thickening of the pericardium confirmed on CT or MRI; (2) restrictive mitral inflow pattern on echocardiography and/or pulmonary venous inflow; and (3) the absence of significant respiratory variation of early diastolic filling velocity (<25% mitral E velocity variation). Webtable 1 shows the underlying aetiology of 38 patients with mixed constriction and restriction confirmed on cardiac catheterisation and, in some cases, surgical findings.[10] Just under half (45%) of the patients underwent pericardial stripping and none were transplanted. Overall 5 and 10 year survival rates were 60 (8)% and 50 (10)%, respectively.[10] Survival appears worse in the mixed constriction and restriction group than with constriction alone.[10] Of the patients with mixed physiology, there was no significant difference in survival between the surgically treated group and the medically treated group.[10] Treatment options in this complex group include medical therapy, pericardectomy and heart transplantation in selected patients.

## CONCLUSIONS

Pericardial disease encompasses a complex spectrum of disorders. Correct management requires an integrated approach including sound clinical skills and a good knowledge of cardiac physiology. In addition, skill in the interpretation of a variety of imaging modalities and haemodynamic data is required. Cardiologists should have the skills to perform a needle pericardiocentesis and an awareness of the possibilities and limitations of pericardial surgery. As the understanding of pericardial disease evolves, cardiologists must apply the evidence from clinical studies to individual patients to ensure the best possible outcomes in patient care.

### Echocardiographically guided pericardiocentesis: key points

In all but emergency cases, echocardiographic guided pericardiocentesis should be performed in an area where the patient can be closely monitored.

Continuous ECG monitoring, invasive or 2 min non-invasive blood pressure monitoring, and continuous pulse oximetry should all be available.

The best technique in an individual patient depends on the amount and location of the effusion, the patient's clinical status and the operator's experience.

Surgical treatment should be performed in patients with pericardial effusions caused by aortic dissection, a direct puncture wound or ruptured ventricular aneurysm. The steps we employ for non-emergency echocardiographically guided percutaneous pericardiocentesis are summarised below:

- The patient's echocardiogram is reviewed. The approach is chosen where the most direct route to the maximum amount of pericardial fluid occurs while avoiding any vital structures.
- Laboratory tests are ordered or reviewed. Any coagulopathy, electrolyte disturbance (especially hypokalaemia) or severe anaemia should be corrected.
- Intravenous access is obtained and $O_2$ is delivered to maintain oxygen saturation of >90%.
- Echocardiography is performed by the bedside. A proposed entry point is marked on the patient's skin where the percutaneous needle should penetrate the chest wall, and the transducer angle is noted as this will need to be replicated by the pericardiocentesis needle. The distance from the chest wall to the effusion and the distance to the nearest cardiac structure is determined as this will determine the maximum distance that the pericardial needle can be safely advanced. If an acoustic window is available, remote to the proposed puncture site, needle puncture can be directly visualised. If not, the imaging probe should be covered with a sterile cover, and available for the physician performing the procedure to use if needed.
- Local anaesthetic is infiltrated. The pericardiocentesis needle is inserted and advanced along the previously determined pathway. Constant aspiration during needle advancement followed by a slight pause for injection should be performed to make sure that a plug of tissue or clot is not occluding the needle tip.
- Once the pericardium is entered, fluid should flow freely into the syringe. If there is any doubt, the position of the needle can be confirmed by injecting agitated saline contrast. A guidewire is introduced into the pericardial space and its correct position can usually be verified echocardiographically. Dilators are used to enlarge the guidewire track. A pigtail catheter is thread over the wire into the pericardial space. The pericardial effusion should be drained completely, if possible. Intermittent echocardiographic re-evaluation of the pericardial fluid volume throughout the aspiration procedure is useful to assess the degree of remaining fluid.

The pericardial catheter is usually left in situ for 12–36 h. Once drainage ceases, the patient is re-imaged and the catheter is removed when only a minimal amount of fluid remains.

Additional references appear on the *Heart* website— http://heart.bmj.com/supplemental

## INTERACTIVE MULTIPLE CHOICE QUESTIONS

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In compliance with EBAC/EACCME guidelines, all authors participating in Education in *Heart* have disclosed potential conflicts of interest that might cause a bias in the article. Brad Munt has received grants from the Heart and Stroke Foundation British Columbia and the Yukon and the Diamond Foundation

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